Pregnancy Management of Women with Coronary Artery Aneurysm Due to Kawasaki Disease During Childhood: Case Report and Review of the Literature

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Abstract

Kawasaki disease (KD) is an acute systemic vasculitis occurring during childhood. It could be associated with persistent heart defects such as coronary artery aneurysms (CAA). During pregnancy, there is a theoretical high risk of full myocardial infarction and sudden death. Guidelines do not exist but case report studies found in the literature show that these pregnancies could have a good maternal and fetal outcome dependent upon a careful follow pre-conceptional counselling is important for evaluating thecardiological risk factors. Maternal cardiovascular complications are rare when anticoagulation therapy is given. Low dose aspirin is safe and prescribed to patients with coronary aneurysms. For the management of labour and delivery, assisted second stage of labour with locoregional anesthesia is recommended. C-sections should be offered only to patients with severe cardiological conditions or for obstetrical reasons. In conclusion, multidisciplinary team management of pregnant patients with KD sequelae remains a challenge but is essential for a favourable outcome of the pregnancy.

Long-term outcome of adults and also pregnant women with previous history of KD is now being evaluated. Therapies such as angioplasty and cardiac transplantation may be necessary but in mild cases, only prevention of coronary thrombosis is needed. Generally, patients are treated with a low dose of aspirin [6]. Depending on the size and the age of the patient, antiplatelet agents can be added. In giant coronary aneurysms, aspirin is combined with an anticoagulation treatment such as warfarin but this is contra-indicated during pregnancy [7,8]. The aim of our paper is to consider the management of pregnant women with CAA during pregnancy and delivery.

Case Report

A 19-year-old primigravida Caucasian patient, with KD during childhood, was referred to our centre at 33 weeks of gestational age for a perinatal management of the pregnancy. At the age of 4 years old, she was diagnosed and treated for KD. Multiple aneurysms were diagnosed on the coronary arteries, left iliac artery, left humeral artery and right iliac artery. She underwent a triple coronary bypass at the age of 10 for an obliterated aneurysm. The patient has no other risk factors. Pregnancy was allowed but because of cardiovascular sequelae, she was under daily low dose aspirin (100 mg) and monitored once a month by her obstetrician and cardiologist.

Throughout the pregnancy, maternal heart rate and blood pressure remained stable. The electrocardiogram (ECG) was normal with sinus rhythm and adequate atioventricular conduction. Echocardiography showed a mildly dilated left ventricle. Contractility and ejection fraction of the ventricle remained within the normal range. A stress echocardiogram and ECG showed a normal heart function and no sign of ischemia. Haemostasis tests were within normal range.

Fetal outcome was uneventful: morphological scan showed no abnormalities and fetal growth was normal. At 37 weeks, because of a breech position, an external cephalic version was attempted but failed. Therefore, a C-section for obstetrical indication was planned at 38 weeks under epidural anaesthesia. A baby boy was delivered with a normal birth...
weight of 2.770 kg and an Apgar of 8/8/8 at 1, 5 and 10 minutes of life. His outcome was normal.

In order to prevent a postpartum thrombotic event, a prophylactic treatment with low molecular weight heparin (LMWH) was administered for 10 days and replaced by low dose aspirin. Progestogen-only contraception instead of oestroprogestative pill was recommended because of a possible thrombogenic effect.

Maternal echocardiography at 5 days post-partum showed a normal biventricular heart function and a mild but not significant tricuspid insufficiency. No sign of thrombosis was observed.

The patient was discharged at day 6.

Discussion

System dynamic models show that by 2030, 1 per 1600 person in the USA will have a medical history of KD [9]. Therefore, an increasing number of pregnant women with potential cardiovascular anomalies will have to be managed. Although rupture of aneurysms is rarely seen, complications such as coronary stenosis, cardiac infarct and sudden death can be observed during pregnancy [10].

Despite an increased incidence of pregnant women with a history of KD, guidelines about the management are unclear. Only 61 women with cardiovascular complications of KD giving birth have been reported in the literature, affecting mainly Japanese patients.

Women with a history of KD should be seen for a preconception counselling. Echography and stress ECG should be done to assess the remaining cardiovascular anomalies. Assessment of coronary arteries should be available usingangiograms. Even though severe cardiovascular accidents reported in the literature are rare, if necessary, women are encouraged to undergo an MRI at 12 weeks of gestational age to confirm the coronary patency [11]. Blood pressure should be closely monitored as pre-eclampsia has been reported in one case [12].

Because of an increased thrombotic risk associated with pregnancy and the presence of aneurysms with low blood flow, anticoagulation therapy should be discussed. Low dose aspirin (60 mg/day to 80 mg/day) could be administrated throughout pregnancy. According to Sibai et al. [13], aspirin doses of 60mg/day during the pregnancy are safe for the foetus. Tsuda et al. [14,15] studied 40 pregnant patients with CAA due to KD. Half of them (19/40) did not receive any medication. There were no cardiovascular complications. The authors concluded that anticoagulant therapy should be administrated only to selected high risk patients. However, Shear et al. [16] recommend low dose aspirin as a preventive treatment during the second and third trimester of the pregnancy. Hibbard et al. [17] suggest the use of anticoagulation drugs for women with giant coronary aneurysms caused by KD. Gordon et al. [12] used aspirin for all of their patients and anticoagulation for three out of four patients. None of their patients experienced any cardiovascular complication during pregnancy [18]. Mc Andrew et al. [19] reported on the only patient with severe cardiac infarct but this patient also suffered from ventricular fibrillation and placenta praevia. It is, therefore, difficult to identify the cause of the infarct.

Obstetrical complications reported are rare. However, Gordon et al. [12] reported preeclampsia and preterm labour in 1/10 patients, probably associated with a deteriorated maternal cardiac condition and also one case of trisomy. In most of the cases, an assisted vaginal delivery under epidural anesthesia is the best option. Caesarian section is indicated for obstetrical reasons or severe cardiac conditions. During postpartum, LMWH should be prescribed basically for preventing CA thrombosis. One case of postpartum haemorrhage was noted and associated with an early reinstitution of anticoagulant therapy [12]. The need for an anticoagulation treatment such as warfarin is still discussed.

Neonatal outcome was generally uneventful. However, one study highlights an increased prevalence of KD in the offspring, suggesting the need for genetic counselling [12].

Conclusion

Although there are no specific guidelines for the management of pregnancies in women with coronary aneurysms, it is recommended to refer to those for pregnancies with cardiovascular compromise [16].

Antenatal anti coagulation therapy by low dose aspirin is recommended by some authors but is controversial. All studies agree that LMWH should be given during post-partum. However, as a risk of post-partum haemorrhage has been reported, LMWH should not be administered too early after delivery [14,15].

Tsuda et al. [14,15], Hibbard et al. [17], Gordon et al. [16], Hayakawa et al. [20] agree that the mode of delivery should be mainly dependant on the cardiovascular status and obstetrical conditions. Vaginal delivery could be safely chosen in the absence of complications. If the patient has severe cardiovascular symptoms such as heart failure and severe ischemia, termination of pregnancy could be an option in the previability period. Caesarean section should be recommended only for obstetrical indications and severe cardiovascular compromise. It should be noted that it does not prevent heart infarcts during labour [14,15]. Forceps delivery could be offered to avoid maternal stress and tiredness during delivery [15]. Epidural anaesthesia should be offered during labour to keep haemodynamic functions stable [13].

A multidisciplinary team including obstetrician, cardiologist and anesthesiologist is essential for the management of pregnant patients with KD sequellae. These high risk pregnancies remain a challenge but an adequate antenatal and perinatal follow up allow a favourable outcome of the pregnancy in most of the cases.
References